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Acute and Recurrent Pericarditis

Still Idiopathic?

Cremer et al. (1) underlined relevant clinical points and the role of cardiac magnetic resonance in their excellent review. The authors also raised important issues concerning pathogenesis, with which we agree and wish to expand.

The term *idiopathic* pericarditis seems unfortunate in both the first attack and in recurrences and a label of our diagnostic ignorance or unwillingness to search for it. In a biopsy study including 259 patients with large pericardial effusion, the underlying cause was identified by molecular and immunity-histological methods: 12% viral, 35% autoreactive/lymphocytic, 2% bacterial, 15% traumatic, 28% malignant, and 8% other (2). On the other hand, the term idiopathic may be reassuring for the physician but may alarm the patient, who does not understand why all the other diseases, such as hypertension, rheumatoid arthritis, and others, are not idiopathic, whereas their disease is. In acute pericarditis, most cases of idiopathic pericarditis are viral in the first attacks, whereas recurrences are often due to too rapidly tapered drug regimen. Other cases seem immune mediated or autoinflammatory. Possible noninvasive clues for autoimmunity are antinuclear (3) or antiheart (4) antibodies (50% of adults), dry eyes, arthralgias, and a subacute course. Conversely, clues for an autoinflammatory pathogenesis involving a pivotal role for inflammosome are acute attacks followed by complete resolution, strikingly elevated C-reactive protein, high fever, and pleuropulmonary and systemic involvement; these patients are generally antinuclear-antibodies negative. A recent randomized controlled trial showed that anakinra has a spectacular effect in these patients (5), and the term idiopathic seems inappropriate for a disease treated with anti-interleukin-1 agents.

The pathogenesis of pericarditis is now comparable to most other inflammatory diseases, and we may consider abandoning the term idiopathic also in this condition.

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REPLY: Acute and Recurrent Pericarditis

Still Idiopathic?

Dr. Brucato and colleagues raise an important issue in the nomenclature of pericardial disease: diagnoses of most patients with acute or recurrent pericarditis are labeled *idiopathic*. As the authors note, the term idiopathic elicits a sense of complacency among physicians. For patients, the response is the opposite. They feel frustrated and alarmed that their clinicians seem to know so little about their disease. Given this predicament, why has idiopathic persisted in pericarditis, and what should the criteria be to abandon the term?

Conventionally, excluding patients with cardiac injury syndromes and underlying autoimmune disease, most patients' conditions are referred to as

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having viral or idiopathic pericarditis. This odd juxtaposition has reflected that we understand the initial cause, a viral infection, but not much regarding the subsequent course. Specifically, the term *viral* alone is unsatisfactory. Most patients with a viral infection, such as Coxsackievirus disease, do not develop pericarditis. Among patients who do, the severity of disease is highly heterogeneous. In addition, identifying the causative virus is generally not a productive endeavor, given the lack of treatment implications. Moreover, recurrences are usually not heralded by repeated viral infections. Often, they occur due to rapid tapering of anti-inflammatory drugs.

However, as we and others have highlighted (1,2), the understanding of the underlying pathophysiology of pericarditis has recently improved. In particular, certain patients have a diathesis toward an exuberant autoinflammatory response, often initiated by a viral agent. From a practical perspective, however, any change in nomenclature should be relatable to the clinician and patient in terms of management. Fortunately, therapies that target the inflammasome have established (3) and emerging (4) efficacy in this disease. Therefore, we agree with Dr. Brucato and colleagues that it is time to consider abandoning the term idiopathic, and we prefer *autoinflammatory* acute or recurrent pericarditis.

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